SYPHILIS OF THE NERVOUS SYSTEM.[1]

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THE importance of this subject cannot be overrated, especially as all the parts of the nervous system are subject to its influence—the brain and its meninges, the cord, the nerves of special sense and the motor and sensory peripheral nerves. Syphilis of the brain is more frequent than of the other parts of the nervous system, the spinal cord being usually secondarily affected, although in many cases primarily, and the same may be said of the peripheral nerves, with the same reservation that here again we may have a primary affection. In a paper on "Cerebral Syphilis," read before the section on practice of the Academy of Medicine, December 18, 1888, I divided syphilitic disease of the brain into three classes, as laid down by Rumpf in his admirable and exhaustive work on "Syphilis of the Nervous System," viz., those involving the cranial bones, the brain and its meninges, and the cerebral vessels. This division will apply equally well to the spinal cord —syphilitic disease of the vertebral column being, however, much rarer than that of the cranial bones. Oppenheim, in a recent article in the "Berliner klinische Wochenschrift," Nos. 48 and 49, 1889, states that in cerebral syphilis, in the majority of cases, the meninges are involved, and that the base is more often affected than the convexity. We have then a meningitis diffusa basilaris syphilitica. Post-mortem it gives the appearance of a mass of paraffine, involving the intra-peduncular space, Sylvian fissures, optic chiasm and the cranial nerves.

Connective-tissue growths extend into the cortex; but in this situation it is not of so much importance as in other regions, as, for example, the motor areas; the cranial nerves appear gray and swollen, from the infiltration of the new growth into the nerve sheath and later into the nerve fibres. This is especially frequent in the optic and third nerves.

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The arteries are always affected, the adventitia being infiltrated by round cells from the meninges. The intima becomes thickened, and obliteration of the vessels occurs, either by consequent narrowing of the lumen or by thrombosis. Softening and hemorrhages into the brain substance often occur, more often due to endarteritis. In disease of the vessels, independent of meningitis, which very often occurs, the granulation-tissue growth may originate in the intima or media.

The pathology of syphilis of the cord is similar to that just given of the cerebrum. Jürgensen believes it to be almost always secondary—that is, that we have a descending lesion, a cerebro-spinal meningitis. We find usually a diffuse meningitis, although not infrequently a circumscribed form, resembling then, in its symptomatology, a tumor or gumma. Cell infiltration takes place into the cord after following the course of the arteries, so that in many cases it would appear as if the initial lesion was in the neuroglia, with secondary involvement of the nerve tracts. Adamkiewicz, at least, maintains this theory in reference to the primary affection in tabes dorsalis, which is so often specific in origin.

Oppenheim regards cerebro-spinal meningitis syphilitica diffusa as one of the most frequent diseases of the nervous system. In the cord, also, we may have endarteritis independent of meningeal affections. The nerves, as we have said, both cranial and spinal, are very often secondarily involved, but not seldom, and especial, is this so of the oculo-motorius, optic and the trigeminus, and less often of the spinal nerves are they primarily affected, resulting in nutritional changes or neuritis.

We see, from what has been said, that no part of the nervous system escapes from the influence of this all-embracing disease, and in fact the diagnostic point of the disease is its universality and multiplicity of symptoms, marked by disappearance and reappearance in the various stages.

The symptoms of cerebral syphilis are: cephalalgia, which, unless we have a gumma or disease of the bone, is very frequently not localized, but more generally increasing after midnight, arousing the patient from sleep by its intensity; the pain at times extends from ear to ear, like a band over the vertex. Psychical disturbance is often very marked, taking the form of excitement, mania or confusion. Many such cases find their way to asylums. A case of this nature came under my observation, whose history may illustrate it: M., aged forty, traveling agent, was missed by his friends while on a business trip, and was found wandering, in an aimless way, in Chicago; seemed confused and at times delirious; patient was taken home, and two weeks later was placed in an asylum, where his mental condition improved under the iodides. Patient declared he remembered nothing of what had occurred previous to his being placed in the hospital. Ptosis and diplopia developed, the latter disappearing, followed, however, by optic atrophy; later still, spinal symptoms developed, with myelitis, vesical and rectal paralysis and bed-sores. The diagnosis of cerebro-spinal meningitis syphilitica was evident. The history of case showed many periods of improvement, but always a relapse, and finally a steady progress of the disease, despite the most energetic treatment.

Somnolence, very extreme in character—lasting hours, or even days—is not uncommon; and such a condition, associated with intellectual disturbance, loss of memory, apathy and any affection of the third or optic nerves, might be considered almost diagnostic of syphilis.

Syphilis as a cause of general paresis has been much debated in recent literature. There is no doubt that a larger percentage of cases of general paresis give a history of syphilis than the other forms of insanity.

The disease is most often located in the arteries. The morbid anatomy bears a close relation to that of general paralysis non-syphilitica. We find pachymeningitis simple and hemorrhagic, thickening of the walls of the blood-vessels, atrophy of the cortex cells and association fibres. The physical signs of the disease are also similar: tremor of the muscles of the face and tongue, difficulty of articulation, inequality of the pupils, ataxia or lateral sclerosis, etc. But often the mental aspect is very different, in many cases ideas of grandeur and self-sufficiency being absent—in other words, we have general paresis, minus the mental symptoms. I had an opportunity last summer, in the Hospital for Paralytics and Epileptics in London, to see many such cases where there was a history of syphilis, with or without alcoholism or the latter alone. The prognosis also is different. We ascribe two or three years as the course of general paralysis, and always give a fatal prognosis, so that I am inclined to think that cases going on for many years, or indeed recovering more or less completely, are due to syphilis.

At times earlier, or again later than the symptoms already detailed, we may have the cranial nerves affected, and, as we have said, especially the oculo-motor and optic as well as the fifth, seventh and eighth, much more rarely the olfactory.

Ptosis, diplopia, inequality of the pupils, with no history of traumatism, orbital tumor or extraordinary overuse of the eyes, in the great majority of cases, indicates cerebral syphilis. We may have one or all the external muscles involved; the internal rectus is, however, especially prone to this affection—optic neuritis and choked disc, or primary atrophy of the optic nerve, as papillitis syphilitica and retro-bulbar neuritis. The lesion lies in the vessels. The nutrition of the nerves by this infiltration of cells becomes sooner or later affected. The ophthalmoscope reveals the arteries thickened and widened and their lumens narrowed, while the veins are enlarged and distorted, the papilla appearing indistinct and swollen. This is probably always secondary to disease of the vessels in the brain.

Extension of the basal meningitis may involve the chiasma or tractus opticus, causing neuritis or atrophy and hemianopsia. These conditions, however, may follow from other causes, and are not alone diagnostic of syphilis.

Hemiplegia and monoplegias, aphasia, motor and sensory, characterized, as we have said, by their disappearance and return, are due usually to arterial disease, resulting in thrombosis or hemorrhage. The same laws as to prognosis are effective here as with hemorrhage and endarteritis and thrombosis from other diseases.

Syphilitic disease of the vertebræ is comparatively rare, and, as the dura does not form its periosteum, slight disease can progress without involving the cord. Exostosis may cause compression of the cord.

Disease of the meninges is much more frequent, and is more often diffused than circumscribed, and indeed in the latter it would be almost impossible to differentiate clinically between it and any new growth or tumor.

The symptoms are divisible into those affecting the meninges, the cord itself, and finally the motor and sensory nerve roots.

We find usually, then, some rigidity or stiffness of the back, shooting pains in the direction of the sensory nerve roots, boring, severe pain at the seat of the lesion, parasthesia and widespread sensory disturbance, and, if the cord is affected, paralysis, motor and sensory, partial or complete, according to the tracts of the cord affected.

The cord itself may be involved independently of the meninges, probably the infiltration in this case following the arteries, as has been said, and involving the neuroglia primarily, later causing by pressure atrophy of nerve cells or fibres. Thus we may have the various forms of atrophy from disease of the anterior horns, myelitis, or again various systemic diseases, locomotor ataxia, lateral sclerosis, etc. The meningeal lesions would differentiate themselves by the neuralgias in the areas of distribution of the sensory roots and the skin affection, atrophy, even though at the same time atrophy of the muscles was present from the lesion affecting the motor nerve roots. A strong proof of syphilis of the cord would be any accompanying cranial nerve affections or history of previous cerebral symptoms, as often ocular paralysis or other cranial nerve affection precede by years the locomotor ataxia or general paresis.

A case now under my observation would seem to be one of specific meningitis, involving the cord and showing also at present disease of the vertebra, probably secondary to the meningitis.

H., aged about forty-five, has always enjoyed good health up to present illness, which began last fall. I saw patient for first time in October, 1889. Patient complained of pain in lumbar region, shooting pains in legs, some numbness in lower extremities, sense of band around the body at umbilicus and intercostal neuralgia of the left side. Examination showed: Dizziness (slight) on standing with eyes closed, gait ataxic, reflexes exaggerated, parasthesia, some vesical weakness, no wasting of muscles. Diagnosis: Combined sclerosis at that time. Under large doses of iodides patient improved very much; neuralgia about disappeared.

About two months later, on entering his house, found himself unable to walk, and had to be carried into the house. It was found that he could not raise his leg from the bed; no atrophy, parasthesia, reflexes still exaggerated; patient subject to sudden seizures of marked rigidity quite painful in character, also to startings or jerkings of the limbs; pain in back, and girdle sensation about the same. Catheter had at first to be used, but not for a long time.

Patient continued the same, with little or no improvement, until two months later, when caries of the vertebræ was indicated by a marked deformity. The probable result will now be all the symptoms of compression myelitis. That we have had, however, a meningitis, with affection of the posterior and lateral columns, irritation of the sensory nerves, with the resulting intercostal neuralgia, there does not seem to be any doubt.

Before closing this paper I would briefly refer to specific affection of the peripheral nerves, either by extension from some syphilitic lymph gland or from specific infiltration of the nerve itself. The changes in the nerve follow the same course as from other lesions, except that as the tendency of all syphilitic infiltration is toward retrogression without suppuration, the nerve may not be completely destroyed, but may regain, in part or in whole, its function. We have then the various nerve affections, neuralgias and sensory and motor paralyses.

Neuralgia may come on at the very onset of the infection, but is usually one of the symptoms of the tertiary stage The supra-orbital and sciatic nerves are very often affected according to Fournier. An affection of the fifth, unless secondary to brain lesion, is rare, although Hughlings Jackson has reported some cases. Cervical and brachial plexus neuralgias are not frequent, nor those of the lumbar plexus.

The nerves of special sense have already been referred to. The olfactory is rarely affected. The auditory nerve, in hereditary syphilis, may later in the disease become affected.

The motor nerves may be separately affected. It is needless to speak further of the third nerve; the seventh is also rarely primarily affected; the spinal nerves may also be affected. The prognosis in peripheral paralysis is a favorable one.

Hereditary syphilis presents the same lesions found in the acquired disease of the cerebral vessels, with hemorrhage, pachymeningitis hemorrhagica and cranial nerve affections.

The spinal cord seems less often affected, although some cases of tabes have been observed. Infantile spastic hemiplegia, with epilepsy and imbecility, initiated by convulsions, is not rare. The appearance of symptoms is sometimes very late, even up to the sixteenth year. Hutchinson, Fournier and others report a number of such cases. Treatment should be carried out systematically and persistently: the inunction method for four or five weeks, to be followed by the iodides for the same time—this treatment, at least the iodides, to be continued for several years. The benefit in many cases of disease of the vessels can be ocularly demonstrated by the ophthalmoscope, when the retinal vessels can be seen to regain their normal state, the thickening of the walls becoming lessened and the lumen being restored. This probably occurs in the cerebral vessels. Wherever we have the cell infiltration peculiar to syphilis, there is hope of causing its absorption or removal; but naturally, when the symptoms are the result of other changes, induced perhaps by the new growth, the results will not be favorable.

A CASE OF DOUBLE PERIPHERAL FACIAL PARALYSIS.

Martin Krüger (Neurol. Centralbl., July 1, 1890) reports a new case of double facial paralysis which was seen in the clinic of Professor Mendel. A workman, 45 years old, had been employed for several years in a laundry, where he was obliged to work ten hours daily in moist air and wet clothes; he was accustomed to drink brandy and beer freely. In September, 1889, he felt drawing pains first in the left cheek, later in the right; then followed tinnitus aurium, constant secretion of tears, inability to close the eyes, sensitiveness to noises, burning in the mouth, dryness of the throat, and disfigurement of the face, which became stiff and immovable like a mask. The naso-labial fold was obliterated on both sides, the corners of the mouth drawn down, the lower eyelids drooped. The effort to wrinkle the forehead resulted only in two horizontal lines. There was also lagophthalmus, difficulty in chewing and drinking, metallic taste in the mouth; at the same time sensation was everywhere normal. The speech sounded inarticulate, difficult and spluttering, had a nasal tone (through paresis of the palatal muscles), and was accompanied with hissing sounds. Taste was affected only on the anterior two-thirds of the tongue.

From the above symptoms the case was one of peripheral double facial paralysis, with the lesion located within the fallopian canal, probably just above the great superficial petrosal nerve or in the geniculate ganglion itself. Electrical examination gave the reactions of partial degeneration, especially marked in the upper branch of the nerve. For this reason and because of the probable rheumatic origin, the prognosis was rather favorable. Under labile treatment with the negative pole the paralysis was greatly improved.

J. W. B.